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A CASE OF SPINA BIFIDA WITH SUPPURATIVE
SPINAL MENINGITIS AND EPENDYMITIS,
DUE TO BACTERIA ENTERING THE
WALL OF THE SAC.

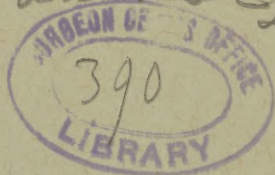
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FIG. 4. Section through the brain cortex, including the wall of the ventricle from the apex of the occipital lobe.

A. Necrotic membrane containing the majority of the cocci.

B. Zone of granulation tissue.

C. White matter.

D. Gray matter.

E. Pia mater showing the exudative meningitis.

The heavily shaded areas indicate the congested blood-vessels.



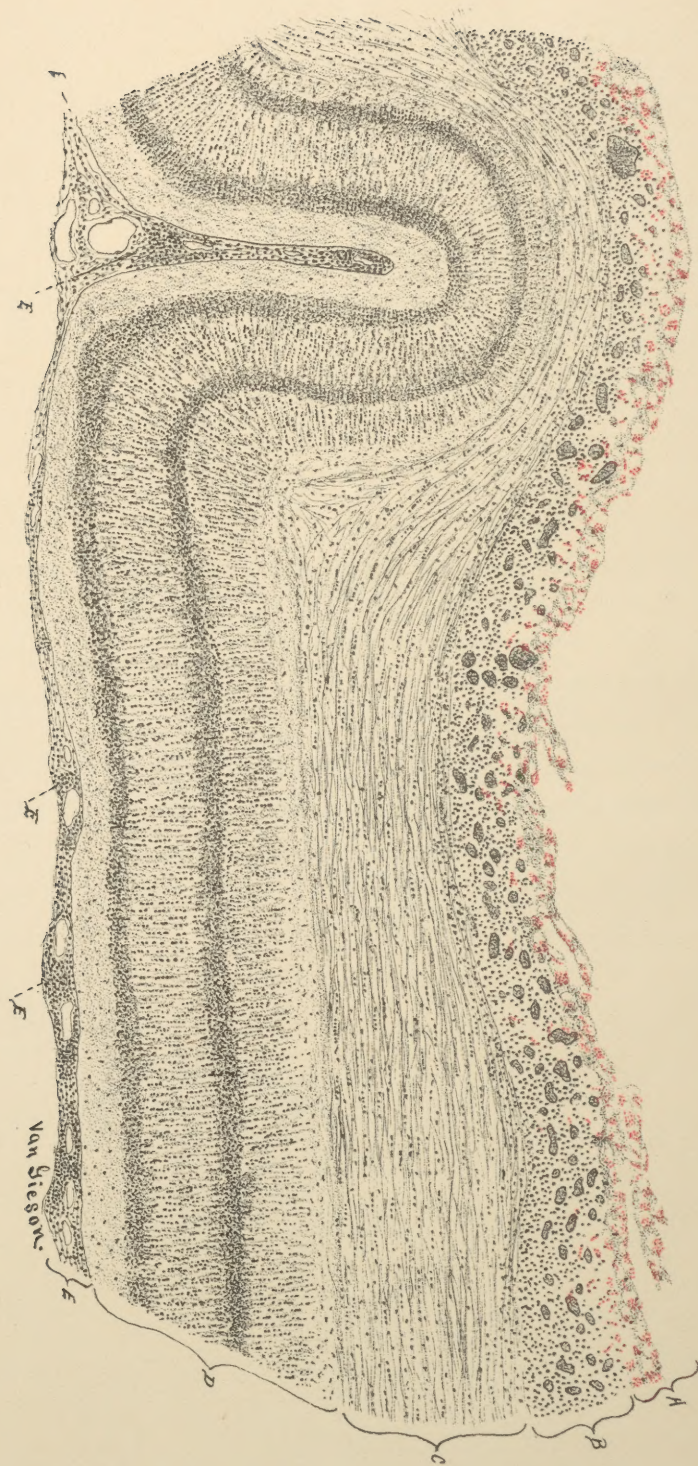


FIG. 3.—Section of the dorsal cord, showing the spinal meningitis, the pellicles on the pia mater, the congestion of the gray matter, the condition of the central canal, and the distribution of the small cocci.

A. Infiltration of the pia mater distending the bottom of the anterior fissure.

B. One of the sulco-commissural vessels passing through the anterior commissure to the margin of the central canal.

C. Canals of the sulco-commissural vessels distended with the meningitis exudation.

V. Spin. ant. and art. spin. ant. anterior spinal vessels. Ven. sept. post. congested vessels of the posterior septum.

P. R. Posterior roots.

X. X. Semi-necrotic pellicle on the posterior surface of the pia mater.

FIG. 2.—A section through the wall of the sac, including one of the minute columns of necrotic tissue extending through the thickness of the skin to its outer surface.

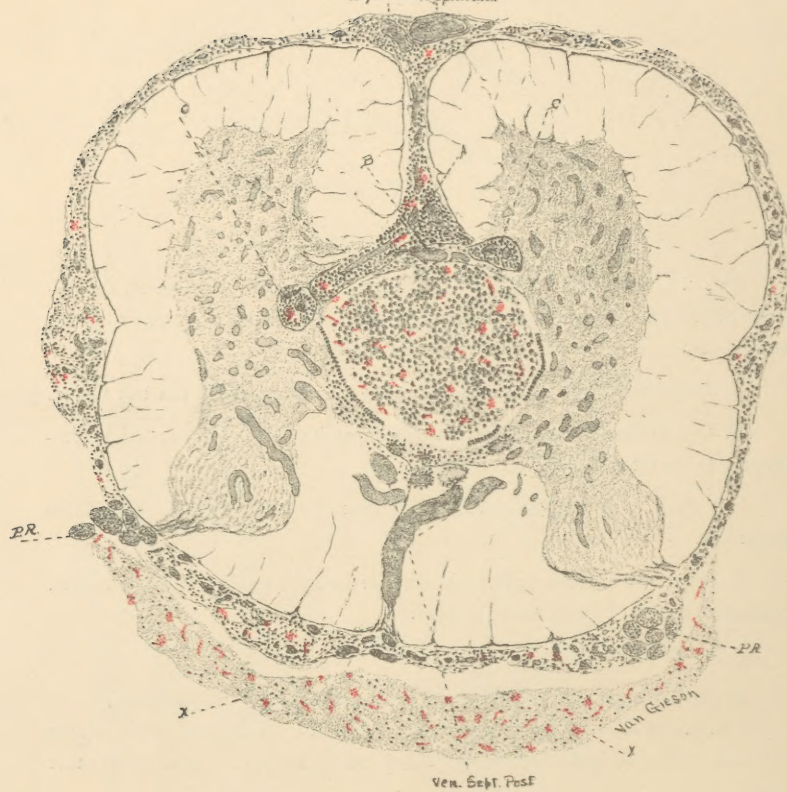
a. Zone of exudation and necrotic matter coating the internal surface of the sac containing the small cocci.

b. Deeper subcutaneous tissue, with congested blood-vessels.

c. Infiltrated corium and papillary derma.

d. Plug-like continuation of the necrosis extending through the skin.

e. Epidermis.



A CASE OF SPINA BIFIDA WITH SUPPURATIVE
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DUE TO BACTERIA ENTERING THE
WALL OF THE SAC.¹

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IN some few cases of spina bifida a suppurative inflammation of the wall of the sac occurs, and leads to a severe and fatal inflammation of the meninges or ventricles of the central nervous system. This complication has attended operations on the sac often enough to be spoken of in the text-books as one of the important contra-indications to punctures or other operations on the wall of the sac. The text-books allude to this complication as purulent arachnitis or purulent meningitis. The percentage of cases of spina bifida in which this complication occurs, or how often the suppurative inflammations of the central nervous system are associated with a like condition of the wall of the sac, is not stated precisely in the available statistics of spina bifida.

The case presented in this paper (which was under the charge of Dr. Holt) shows quite clearly how such a purulent inflammation of the wall of the sac and its dependent lesions of the central nervous system are due to the entrance

¹ Read before the New York Neurological Society, Nov. 4, 1890.

of pyogenic bacteria into the sac, and to their extension along the fluids, meninges, or central canal of the spinal cord, to the brain meninges and ventricles.

The skin of the spinal sac, in spina bifida cases, would seem especially permeable for bacteria. It is frequently extremely thin and the central cicatrix has occasionally a raw exposed surface and lacks the epidermis covering. In surgical operations, pyogenic bacteria have probably been introduced into the sac in the cases where purulent central neural inflammations or suppurative inflammation of sac-wall, or both of these, have followed the operation.

Clinical History (by Dr. Holt).—A female infant, three days old, was admitted to the Babies' Hospital, March 2, 1890. The family history was negative so far as other deformities were concerned. On admission the child was plump and well nourished and presented no physical signs of hydrocephalus, the head being normal in size and the fontanelles not bulging nor tense. There was a lumbosacral tumor, measuring $3\frac{3}{4}$ by 6 cm. in diameter and projecting $1\frac{1}{2}$ cm. above the spine. Its walls were exceedingly thin, so that translucency was almost perfect. The skin covering the tumor was healthy except over a central elliptical space—the central cicatrix—directed vertically in the median line, about 1 cm. wide and $2\frac{1}{2}$ cm. long. This extent of the skin, corresponding to the central cicatrix, had the appearance of a granulating surface, and was covered with a sero-purulent discharge. Neither cord nor nerve-filaments could be made out in the sac.

The feet were in the position of ordinary typical equinovarus of rather an extreme degree. The test of power in the lower extremities was rather unsatisfactory. There did not appear to be any incontinence of fæces, but the condition of the bladder could not be determined with certainty. There was a small umbilical hernia.

The temperature was taken for the first two days, but, as it remained normal, the record of the temperature was then discontinued. The tumor was covered with sub-iodide of bismuth; over this was laid a compress covered with vaseline, and over this a large pad of absorbent cotton held,

in place with a flannel roller applied so as to make gentle uniform pressure over the sac.

During the first two weeks the child was quiet and slept most of the time, and presented nothing abnormal in the way of symptoms. At the end of this time there was no longer any doubt regarding paraplegia. It was nearly complete; the thighs were habitually slightly flexed, while the legs were extended. Sensation was still doubtful. There was no incontinence of fæces, and retention or incontinence of urine was apparently absent.

At this date the tumor was noted to be perceptibly smaller, its surface somewhat wrinkled, and its translucency was very imperfect. The wall of the sac seemed to be thickened. On account of these changes in the sac, it was decided to postpone any operative procedures.

During the third week the general nutrition failed quite rapidly. The infant screamed much of the time and was quite restless. The temperature record was not kept, but it did not seem to be above the normal. There was no opisthotonos or vomiting; the bowels were regular and food was taken regularly.

Early in the morning of March 21st the infant was found cyanotic, with a very feeble pulse, and it died quietly a few hours later. There were essentially no symptoms during the last week, except those of rather marked irritability and failing nutrition.

AUTOPSY (three and a half hours after death).—There was nothing abnormal about the thoracic or abdominal viscera. Head of normal size; the fontanelles not bulging; slight overlapping of the cranial bones.

Brain.—Five ounces of rather thin yellow pus escaped from the lateral ventricles while the brain was being taken out. The whole cerebrum was rather soft; the convolutions were not markedly flattened. The pia mater of the convexity seemed normal. Its vessels were filled, but not distended, and there were no evidences of meningitis. The sinuses were free of thrombi. At the base of the brain there was quite a thick ($\frac{1}{2}$ mm.) yellowish pellicle in the median

line of the anterior surface of the medulla, which spread out in a thin layer, on either side, over the adjacent surface of the cerebellum, about one-half of which was thus coated. There was a similar pellicle on the optic tracts, about the optic commissure, and over a small area of the median anterior surface of the pons.

The Ventricles.—The lateral ventricles were very greatly dilated, measuring together $8 \times 6 \times 3\frac{1}{2}$ cm. in diameter, while in the normal brain of a child of the same age they measure together about $\frac{1}{2} \times 5 \times 2$ cm. The volume of the lateral ventricles in this case must have been at least forty or fifty times their volume in a normal brain of the same age.

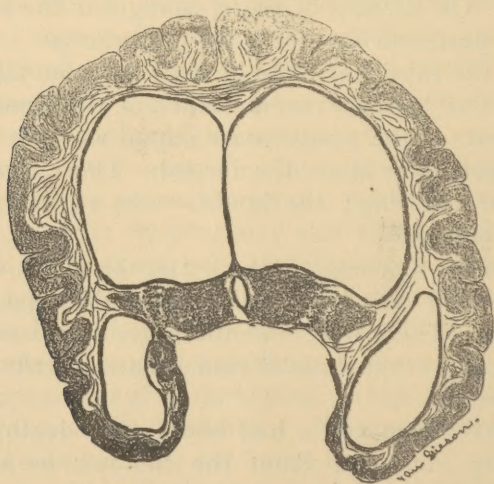


FIG. 1. —Transverse vertical section through the brain at the junction of the anterior $\frac{3}{8}$ and posterior $\frac{2}{8}$. The shaded zone lining the ventricles indicates the zone of granulation tissue—about $\frac{2}{3}$ of natural size?

In the section of the brain, shown in Fig. 1, the posterior horns measured 3 cm. in diameter. While the ventricles are thus enormously dilated, there is no corresponding expansion in diameters of the brain; the outer surface of the brain is not increased: the brain itself was normal in size, or perhaps a trifle smaller than usual. The increase of the volume of

the ventricles is formed apparently at the expense of the subjacent surrounding brain-tissue or by a condensation of the latter. The convolutions with their white matter are in places reduced to a mere shell in thickness (at the posterior and descending horns), measuring but 2 to 3 mm., about one-tenth of the thickness of the corresponding portion of the normal brain at the same age. The average thickness of the cortex is about 8 mm. The basal ganglia are only about half their normal vertical diameter and are soft. The lateral ventricles and their horns and the third ventricle are everywhere lined with a thin (1 mm. in diameter) translucent red membrane, having the appearance of granulation-tissue, which can be readily stripped up from the subjacent brain-tissue. Lying over this reddish membrane in many places are extensive flocculi of yellowish white material. The fourth ventricle is normal, except that its floor is congested. The medulla is much congested.

The meninges and substance of the *spinal cord* are uniformly congested. Lying on the surface of the pia mater are scattered yellowish white pellicles, which are most extensive at the lower part of the cord.

The *sac* contained about half an ounce of pus, and its walls were thickened by a thick (2 to 5 mm.) yellowish white layer lining its internal surface. The cauda equina entered the sac and passed along its walls in the usual way.

MICROSCOPICAL EXAMINATION (by Dr. van Gieson).—*The Wall of the Sac*: With the exception of a few scattered heaps of necrotic cells of the Rete Malpighii, the epidermis is absent from the oval central cicatrix of the sac (Fig. 2, *E—E*). In this central portion the papillary and more superficial portions of the derma are densely crowded with small round cells. The subcutaneous connective tissue is also infiltrated, but less extensively, with small round and elongated cells (Fig. 2, *b—c*). Both of these layers are much congested, the subcutaneous connective tissue containing the larger and more irregular vessels.

Beneath or internal to the subcutaneous connective tissue of the skin of the sac is a zone (Fig. 2, *a*) of irregular thickness, with a ragged free internal border, composed of frag-

mentary nuclei, necrotic material, and a scanty amount of fibrin lying on or near the free margin. In this zone the strata near the free margin are more distinctly necrotic than the deepest laminæ bordering the subcutaneous connective tissue; these have the appearance of granulation-tissue in a somewhat necrotic condition. This zone contains great numbers of bacteria of three kinds: a few short, thick bacilli, some large cocci, *and the most numerous predominating form—small cocci in chains and clusters.* The bacteria are most numerous near the free margin of the semi-necrotic zone (Fig. 2, *a*). This is the zone which appeared at the autopsy as the yellowish shreddy membrane coating the inner surface of the sac.

At the autopsy no apertures or solutions of continuity were noticed in the skin of the sac, excepting the excoriation of its central portion; but in the sections from this region in two places, microscopic plug-like continuations of the necrotic zone extend through the entire thickness of the excoriated central portion of the skin of the sac and communicate with the outer surface of the skin, as shown in Fig. 2, *d*. These two minute plugs of necrotic tissue extending through the thickness of the skin contain many of the small cocci, which are also attached to the outer surface of the plugs, and to the outer surface of the skin bordering on the plugs (Fig. 2, *d*).

At the edges of the excoriated central portion of the sac, the epidermis is present and the inflammatory changes are less intense, but the necrotic membrane with its bacteria continues over the whole inner surface of the sac wall.

The Central Nervous System.—The inner surface of the *dura mater spinalis*, in places in the dorsal and lumbosacral regions, is coated with a thin layer of red blood-cells with a little fibrin. The *pia mater spinalis* has congested vessels, and is infiltrated to a moderate degree with small round cells. The *pia mater* passing into the anterior fissure is similarly very abundantly infiltrated (except in the upper cervical region) so that the cell exudation quite uniformly distends the bottom of the anterior fissure (Fig. 3, *A*), and fills up the two large canals in the anterior com-

missure on either side of the central canal (Fig. 3, *c. c.*). These two canals in the anterior commissure, through which the sulco-commissural branches of the anterior spinal vessels pass obliquely into the gray matter, lies so close to the dilated central canal, as to furnish a passageway for the exudation in the anterior fissure into the central canal (Fig. 3).

In several places in the lumbo-sacral region the exudation in the anterior fissure and in the pervascular spaces communicating with the fissure was so abundant as to have broken through the anterior commissure into the enlarged central canal.

As in the anterior fissure, the congested vessels of the posterior septum, in places, are surrounded by investments of small round cells, which are continuous with similar cells in the central canal (Fig. 3). There are scattered pellicles of partially necrotic material upon the surface of the pia mater which are most extensive and form an almost continuous layer upon the posterior pial surface except in the upper cervical region.

The vessels of the *spinal cord* are greatly congested, especially in the gray matter. The *central canal* is large, measuring, in general, about 1 mm. in diameter. It is filled up with leucocytes, necrotic or granular material, and some fluid and fat globules (Fig. 3). A good deal of the epithelium of the central canal has been desquamated or destroyed. There is hardly any evidence of inflammation of the substance of the spinal cord, excepting some perivascular heaps of leucocytes just about the margin of the central canal, and these changes in the wall of the central canal and its contents seem to be due for the most part to the extension of the spinal meningitis exudation through the anterior commissure, and along the paths of the vessels of the anterior fissure and posterior septum.

In the upper cervical region the central canal is normal, the congestion of the gray matter is less intense, and the spinal meningitis less in degree, and in places almost entirely absent. Just before entering the fourth ventricle

the central canal is again dilated and filled up with necrotic and exudative products.

In sections stained with Gram's method, and examined with the Zeiss $\frac{1}{12}$ oil immersion lens, small cocci, like the predominant form in the sac wall, are present in the pia mater, in the anterior fissure and its communicating channels in the anterior commissure, in the exudation plugging the central canal, and in the pellicles lying over the pia mater (Fig. 3).

The *flake or pellicle of pus on the pons* described in the autopsy notes contains small cocci like those in the cord and sac wall.

The walls of the *third ventricle*, the *upper portion of the aqueduct of Sylvius* and of the *lateral ventricles* are everywhere uniformly changed. They are lined by a zone of granulation tissue, bordered internally by ragged necrotic fibrinous layer of variable thickness containing very great numbers of the small cocci similar to those in the cord and sac wall (Fig. 6, *A and B*).

This fibrinous necrotic zone corresponds to the yellowish pellicle, and the granulation tissue zone to the reddish membrane subjacent to the pellicle mentioned above in the gross description of the ventricles. The vessels of the choroid plexus of the lateral ventricles are congested and their meshes filled with small round cells. The *fourth ventricle* is normal, except that the choroid plexus vessels are congested, and there is some proliferation of the choroidal epithelium.

The *pia mater of the convexity* shows in most places a moderate degree of exudative meningitis.

Anatomical Diagnosis.—Spina bifida with hydromyelia. Suppurative inflammation of the wall of the sac. Purulent spinal meningitis, with the exudation extending into the central canal. Suppurative ependymitis of the lateral and third ventricles, exudative cerebral meningitis.

Remarks.—The bacteria were, unfortunately, not identified by culture, but the small cocci are presumably some form of pyogenic bacteria, which entered the exposed surface of the skin of the sac, producing an inflammation of

the wall, and extending along the central nervous system, excited the lesions in the spinal cord and brain.

The *absence* of the *physical signs* of congenital *hydrocephalus* is remarkable. Yet the greater part of the fluid accumulation within the ventricles appears to have been formed at birth, and does not seem to have been caused by the suppurative ependymitis. For in this case the large intra-ventricular collection would have been formed so rapidly—within three weeks—that apparently it would have necessarily produced enlargement of the head and distention of the sutures or fontanelles. Probably at birth the ventricles were nearly as large as at the autopsy, and were filled with clear fluid; then after the lodgment of the cocci in the walls of the ventricles, the exudative purulent ependymitis occurred, which increased the intra-ventricular contents somewhat and converted the clear fluid of the ventricles into a sero-purulent fluid.

The morphological identity of the small cocci in the different regions and the absence in the brain and cord of the other two forms of bacteria in the sac, indicate the small cocci as the real pathogenic agents and that the associated bacteria are merely contaminating forms. As far as can be determined from their dimensions and grouping, and by comparing them with other known species of pyogenic micro-cocci in control sections, the small cocci in this case are thought to be the *streptococci pyogenes*.

The plugs of necrotic tissue extending through the skin of the sac are rather suggestive as to the probable points of entrance of the cocci.

A very interesting feature of the case bearing on the pathology of the hydro-myelia and syringo-myelia is the demonstration of how readily an exudation of the pia mater spinalis in the anterior fissure may communicate with an imperfectly closed central canal, by following the perivascular spaces of the vessels entering the cord, or by breaking through the anterior commissure.

